
بسم الله الرحمن الرحيم

D / AMAT

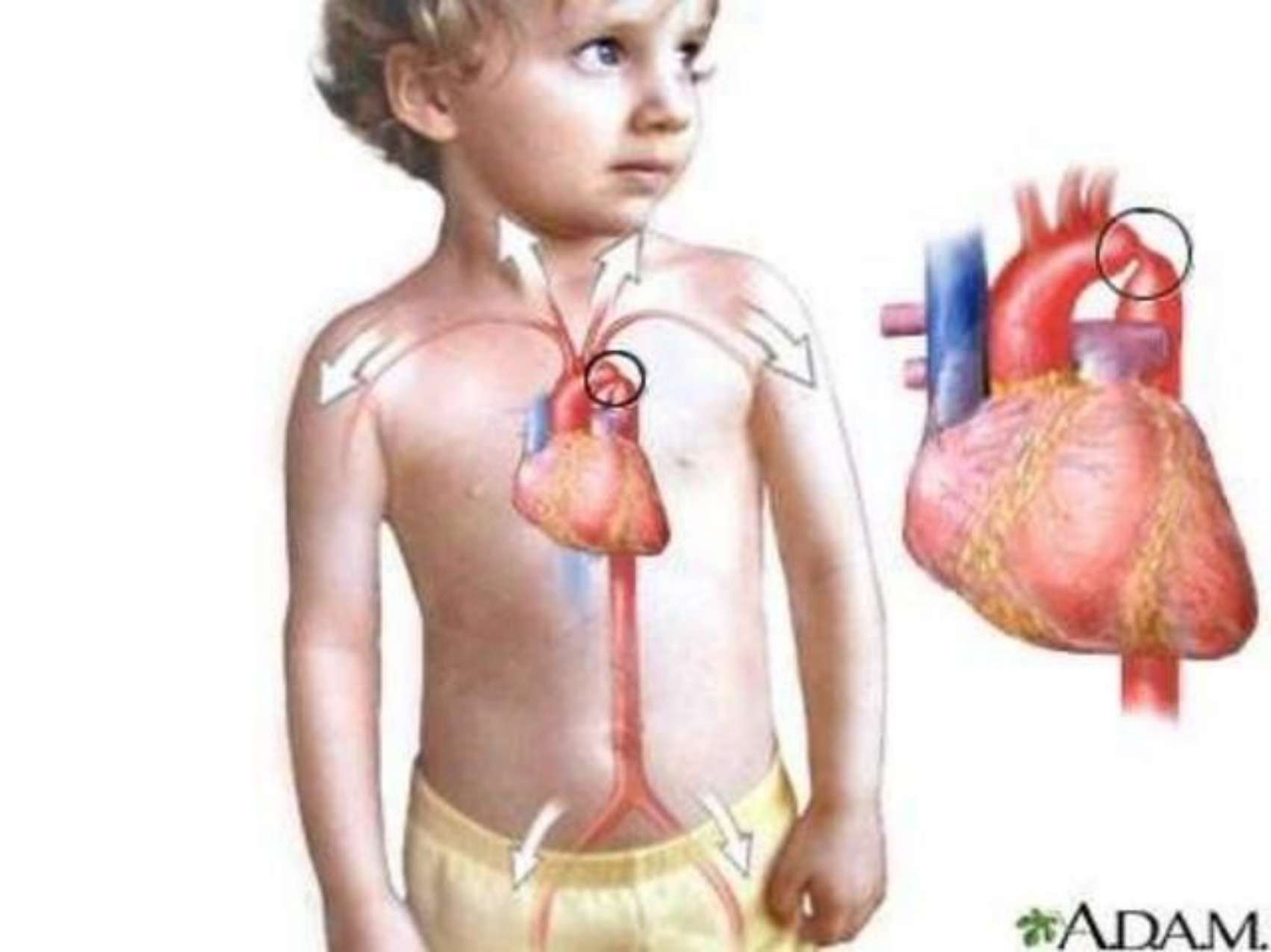
COARCTATION OF AORTA

OVERVIEW

- Definition
- Pathophysiology
- Classification.
- Diagnosis
- Treatment
- Post coartectomy syndrome
- Prognosis.
- Coarctation with VSD
- Coarctation with other cong; heart diseases.

Definition.

- Coarctation of the aorta describes the narrowing of the aorta.
- Can occur at any point from the transverse arch to the iliac bifurcation.
- 98% occur just below the origin of the left subclavian artery at the origin of the ductus arteriosus (juxtaductal coarctation).

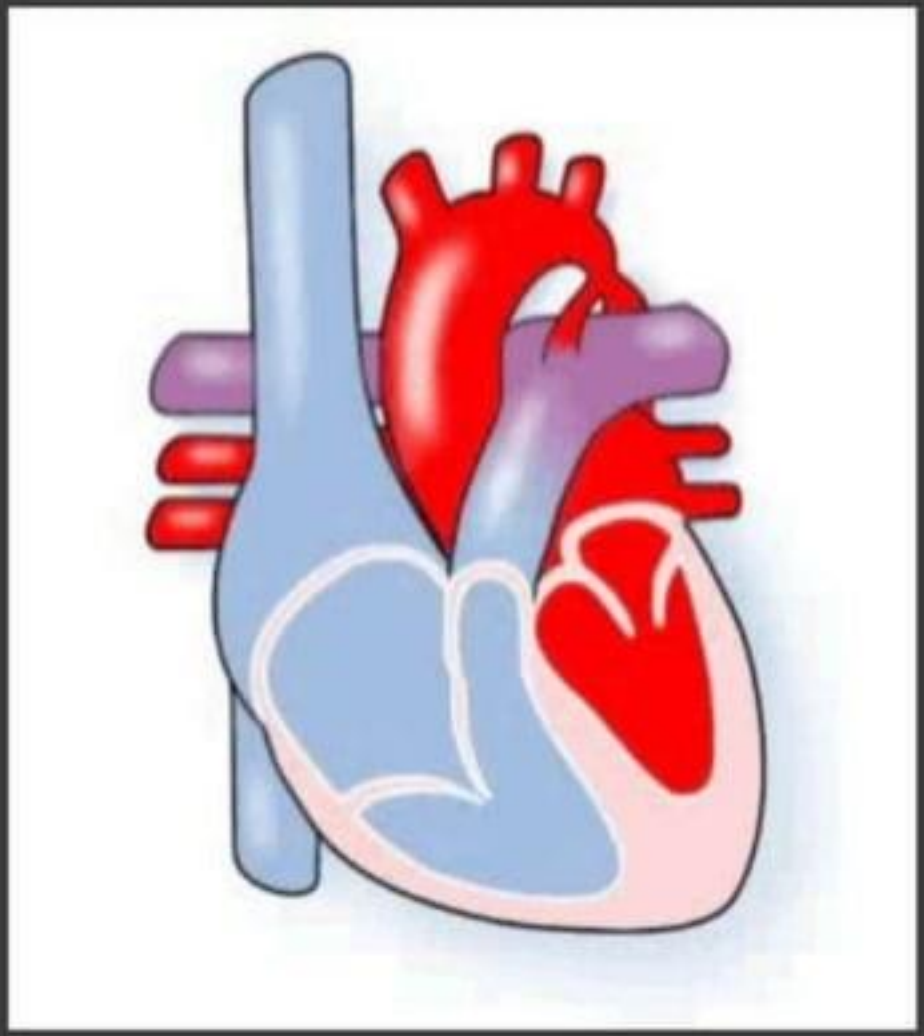

A.D.A.M.

Pathophysiology

- Can occur as a discrete juxtaductal obstruction or as tubular hypoplasia of the transverse aorta, or due to both.

- Alternatively, coarctation may be caused by abnormal extension of contractile ductal tissue into the aortic wall.

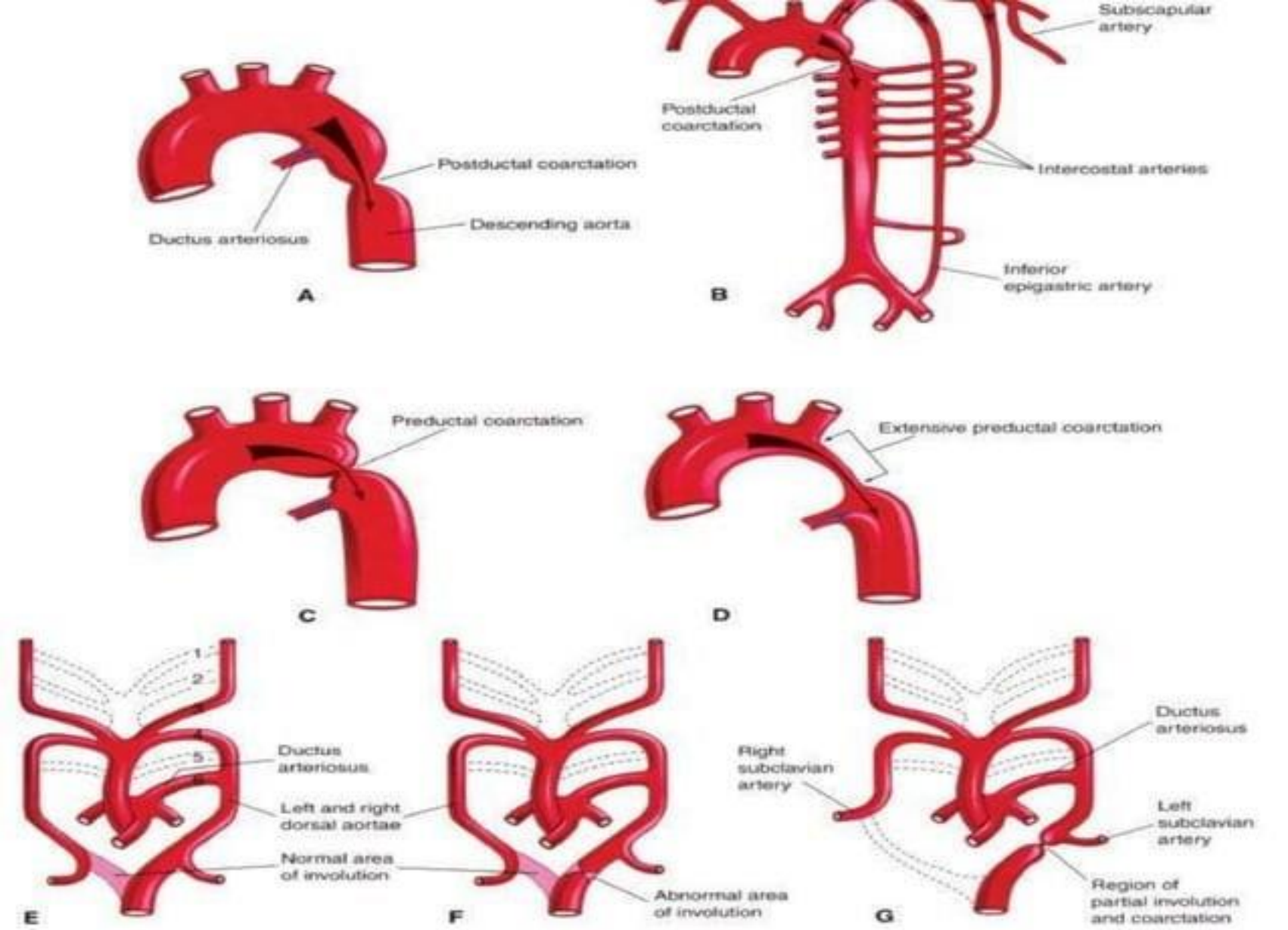
Pathophysiology



In patients with discrete juxtaductal coarctation, ascending aortic blood flows through the narrowed segment to reach the descending aorta, left ventricular hypertension and hypertrophy result.

PATHOPHYSIOLOGY

- In the 1st few days of life, the PDA may serve to widen the juxtaductal area of the aorta and provide temporary relief from the obstruction.
- With more-severe juxtaductal coarctation or in the presence of transvers arch hypoplasia, right ventricular blood is ejected through the ductus to supply the descending aorta
- Unless operated on in infancy, coarctation of the aorta usually results in the development of an extensive collateral circulation



From: Moore, K. L., & Persaud, T. (2008). *The developing human--clinically oriented embryology* (8th ed., p. 323).

PATHOPHYSIOLOGY

- Perfusion of the lower part of the body is then dependent on right ventricular output. In this situation, the femoral pulses are palpable, and differential blood pressures may not be helpful in making the diagnosis
- Ductal R-L shunt manifested in the form of differential cyanosis

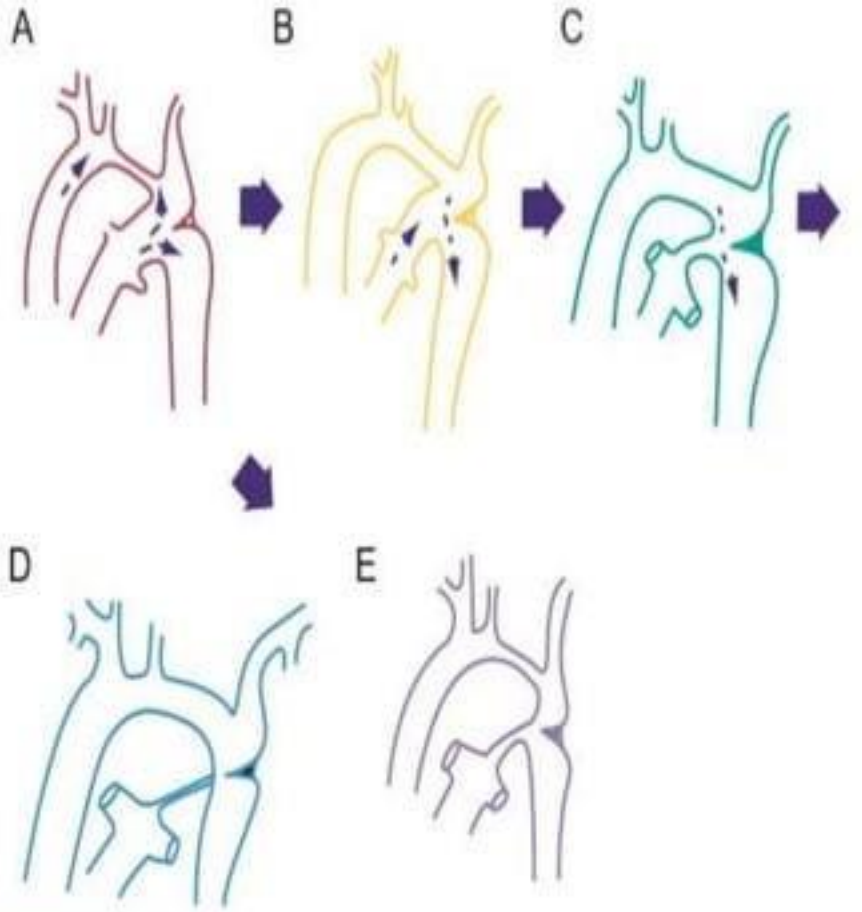


Figure 427-7 Metamorphosis of coarctation. **A,** Fetal prototype with no flow obstruction. **B,** Late gestation. The aortic ventricle increases its output and dilates the hypoplastic segment. Antegrade aortic flow bypasses the shelf via the ductal orifice. **C,** Neonate. Ductal constriction initiates the obstruction by removing the bypass and increasing antegrade arch flow. **D,** Mature juxtaductal stenosis. The bypass is completely obliterated, and intimal hypoplasia on the edge of the shelf is aggravating the stenosis. Collaterals develop. **E,** Persistence of the infantile type fetal prototype. An intracardiac left-sided heart obstruc-

PATHOPHYSIOLOGY

- Infantile type: Coarctation associated with arch hypoplasia, recognized early in infancy.
- Adult type: isolated juxtaductal coarctation, which, if mild, was not usually recognized until later childhood
- B.P is elevated in the vessels that arise proximal to the coarctation; BP as well as pulse pressure is lower below the constriction.

CLINICAL FEATURES.

- Asymptomatic if recognized after infancy.
- Weakness, leg pain after exercise in older children
- Disparity in pulsation and blood pressure in the arms and legs
- The femoral, popliteal, posterior tibial, and dorsalis pedis pulses are weak (or absent in up to 40% of patients), in contrast to the bounding pulses of the arms and carotid vessels.
- Palpate radial and femoral pulses simultaneously for the presence of a radial-femoral delay

Clinical features

- In coarctation of the aorta, blood pressure in the legs is lower than that in the arms

Early presentation

- Young patients may present in the first few weeks of life with;
  - Poor feeding
  - Tachypnea
  - Lethargy
  - Progression to overt CHF
  - Shock.
- These patients may have appeared well prior to hospital discharge, and deterioration coincides with closure of the patent ductus arteriosus.

Physical findings in early presentation

- Tachypnea
- Tachycardia
- increased work of breathing
- Shock
- Blood pressue discrepancies between the upper and lower extremities
- reduced or absent lower extremity pulses to palpation

- However, when the infant is in severe heart failure, all pulses are diminished.
- Upon treatment for heart failure, prominent brachial pulses with weak or nonpalpable femoral arterial pulses may be discerned.

Murmur

- The murmur associated with coarctation of the aorta may be nonspecific initially and is usually a systolic murmur in the left infraclavicular area and under the left scapula.

- Additional murmurs that result from the presence of associated abnormalities, such as VSD or aortic valve stenosis, may also be detected.

- An ejection click may signify the presence of a bicuspid aortic valve, whereas a gallop rhythm may indicate ventricular dysfunction.

Late presentation

- Patients often present after the neonatal period with hypertension or a murmur.
- These patients often have not developed overt CHF because of the presence of arterial collateral vessels.
- Diagnosis is often made after hypertension is noted as an incidental finding during evaluation of other problems.

- Other presenting symptoms may include;
  - Headaches
  - chest pain
  - Fatigue
  - life-threatening intracranial hemorrhage.
- True claudication is rare, although an occasional child may experience pain or weakness in the legs.
- Palpation of femoral pulses and measurement of blood pressure during routine examination is necessary to avoid a delay in the diagnosis.

Laboratory investigations

- Laboratory studies in neonatal patients who present in shock include the following:
  - Septic workup includes blood, urine
  - cerebral spinal fluid (CSF) cultures.
  - Electrolyte levels, BUN, creatinine, and glucose concentration.
- Measure arterial blood gases and serum lactate levels.
- Laboratory studies in older patients who present with hypertension include urinalysis, electrolyte levels, BUN, creatinine, and glucose concentrations.

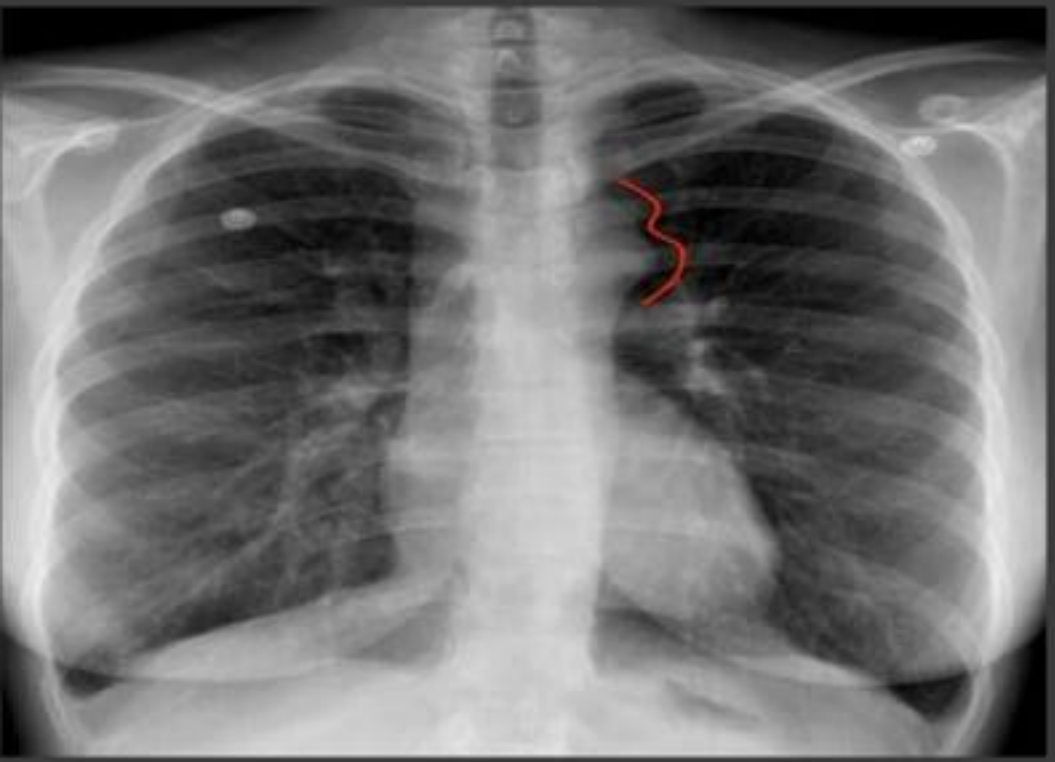
Imaging studies

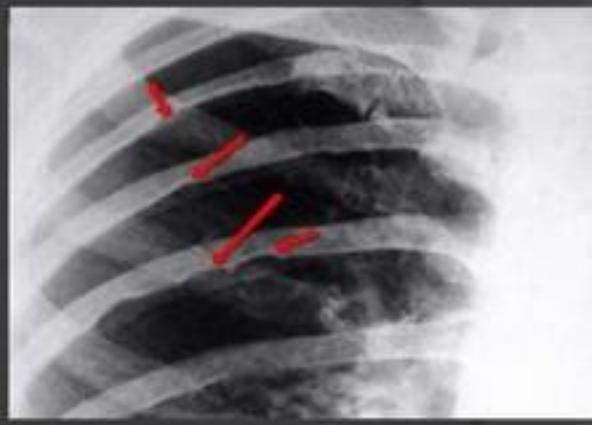
- ***Chest radiography*** in patients with early onset of coarctation of the aorta may reveal cardiomegaly, pulmonary edema, and other signs of congestive heart failure.

- Radiography in patients with late onset of coarctation of the aorta may reveal cardiomegaly.

- An inverted "3" sign of the barium-filled esophagus or a "3" sign on a highly penetrated chest radiograph (frontal view) may be visualized. Rib notching secondary to collateral vessels may also be seen.

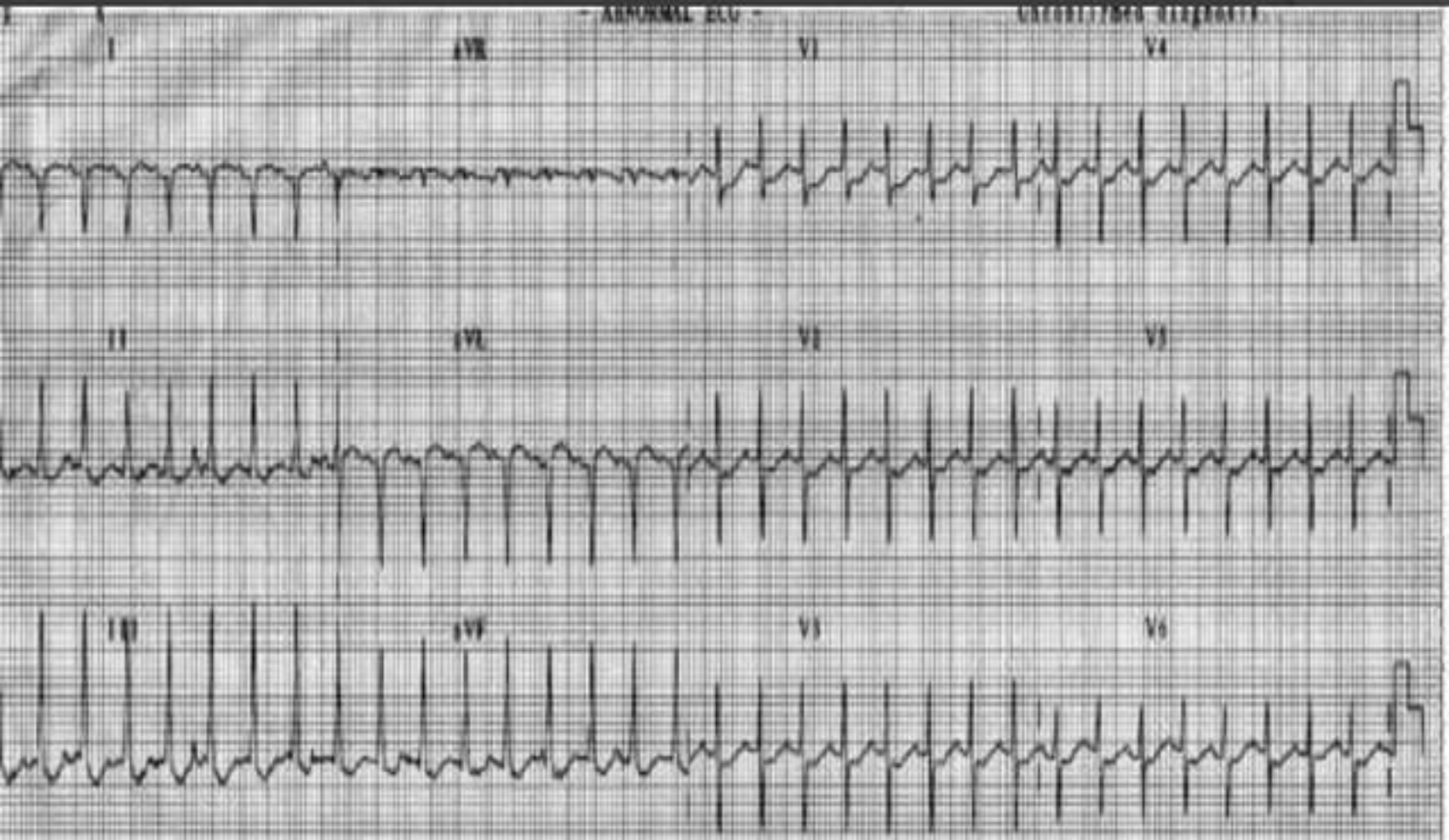
CXR Findings.





Coarctation in infancy

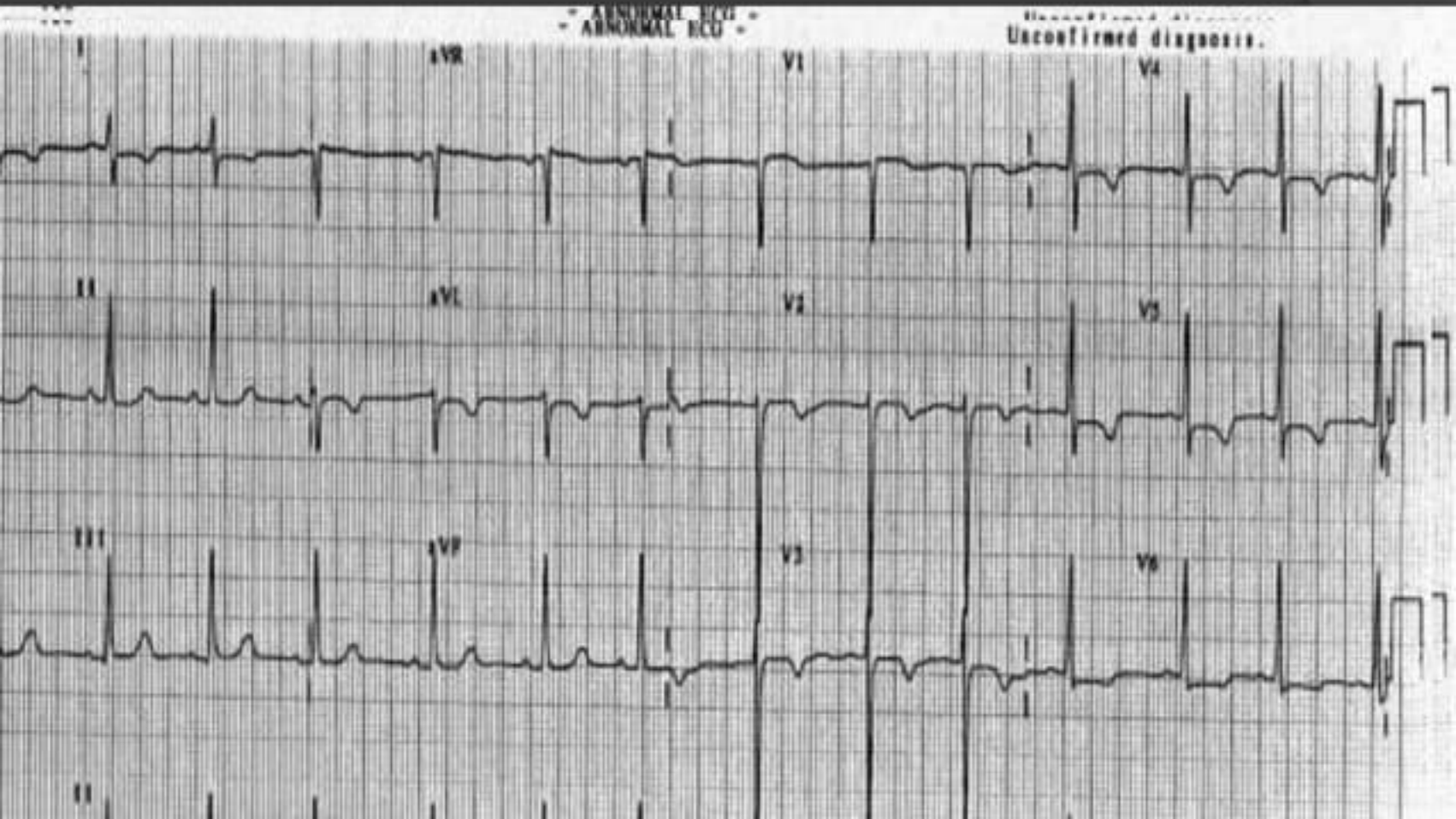
..... Since PAH is common, RV forces are dominant



Coarctation ECG

- In infancy, due to pulmonary hypertension, Right axis and RVH are common
- In older patients, LVH occurs

Coarctation in an older child



INVESTIGATIONS:

- Echocardiography delineates intracardiac anatomy and allows assessment of associated significant intracardiac anomalies

MRI and CT:

- Useful in older or postoperative patients to assess residual arch obstruction, arch hypoplasia, or formation of aneurysms

Investigation:

- Cardiac catheterization with selective left ventriculography and aortography is useful in occasional patients with additional anomalies and as a means of visualizing collateral blood flow.

- In cases that are well defined by echocardiography, CT, or MRI, diagnostic catheterization is not usually required before surgery.

Treatment of early presentation

- Treatment in patients with congestive heart failure (CHF) includes the use of diuretics and inotropic drugs.
- Prostaglandin E1 (0.05-0.15 mcg/kg/min) is infused intravenously to open the ductus arteriosus.
- Ventilatory assistance is provided to patients with markedly increased work of breathing.
- After confirming the diagnosis and stabilizing the patient surgical repair ASAP.

Treatment

- Older children with significant coarctation of the aorta should be treated relatively soon after diagnosis.
- Delay is unwarranted, especially after the 2nd decade of life, when the operation may be less successful because of decreased left ventricular function and degenerative changes in the aortic wall.
- Nevertheless, if cardiac reserve is sufficient, satisfactory repair is possible well into mid-adult life.

Treatment cont:

- Surgical repair:
- Excision and primary anastomosis
- Subclavian flap procedure: which involves division of the left subclavian artery and incorporation of it into the wall of the repaired coarctation has grown out of favor because of a higher degree of residual stenosis
- Patch aortoplasty
- **Primary stent placement:** Newer method.

Post Op complication:

- Rebound hypertension
- Spinal cord injury from aortic cross-clamping if the collaterals are poorly developed,
- Chylothorax,
- Diaphragm injury, and
- Laryngeal nerve injury

POST COARTECTOMY SNDROME

- Acute post period: acute hypertension, abdominal pain
- Anorexia, nausea, vomiting, leukocytosis, intestinal hemorrhage, bowel necrosis, and small bowel obstruction

- Antihypertensive drugs (nitroprusside, esmolol, captopril) and intestinal decompression; surgical exploration is rarely required for bowel obstruction or infarction.

Prognosis:

- Restenosis is rare, children operated before one year of age require repeted surgery
- All patients should be monitored for recoarctation and aneurysm formation
- Ballon angioplasty for recoarctation
- Repair after 2nd decade: Risk of premature CAD
- Severe neurologic disease, SAH, Intracerebral bleed due to cong aneurysm.
- Untreated adult patient succumb between age 20-40 due to HTN encephalopathy, stroke, infective endocarditis, endartritis,

Caorctation with VSD

- Patients with this combination of defects will be recognized either at birth or in the 1st mo of life and often have intractable cardiac failure.
- Presenting ill infant with tachypnea, failure to thrive, and typical findings of heart failure. Often, limb B.P discrepancy is not very marked because of low cardiac output
- Medical management for stabilization of patient; surgery should not be delayed.

Coarctation with VSD

- Many centers repair VSD and Coarctation and at the same time via midline sternotomy.
- Some centers do left lateral thoracotomy with repair of coarctation and pulmonary artery banding in case of complicated VSD.

COARCTATION WITH OTHER CARDIAC ANOMALIES AND INTURREPTED AORTIC ARCH

- HLHS, TGA, Severe mitral or aortic valve disease, and variation of double outlet or single ventricle.
- Coarctation with aortic or mitral valve disease will be treated in context of HLHS.
- DiGeorge syndrome (cardiac defects, abnormal facies, thymic hypoplasia, cleft palate, hypocalcemia). deletion of a segment of chromosome 22q11


THANK YOU